



Case report

Infantile asphyxia due to aberrant uvula – An anatomic misadventure

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ABSTRACT

A case of unexpected death of an infant with an abnormally elongated uvula is presented. The child, born prematurely, was recovering from protracted treatment in hospital, including surgical interventions and periods of ventilation support necessitating recurrent intubations and anesthesia. She was discharged home in good general health, affected by episodes of cough, and was found dead in her crib a week later.

The proximity of the aberrant uvula to the vocal cords may have caused intermittent laryngospasm, with subsequent symptoms of cough and airway obstruction, ending in a fatal outcome. Recurrent airway irritation may have contributed to uvular hypertrophy, due to inflammatory and reactive changes.

It is pertinent for the pathologist to thoroughly examine the structures of the pharynx, and the uvula in particular, in any case of pediatric death suspected to result from asphyxia or sudden infant death syndrome (SIDS).

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1. Introduction

The word 'uvula' is derived from the Latin term "uva", meaning 'little grape', because of its grape-like shape. It is formed in the process of fusion of the soft palate, at eight weeks of gestation. The function of the uvula is not completely understood,¹ but it may facilitate sealing the pharynx during feeding and speech, and has an important role in moistening the oropharyngeal mucosa.²

Since ancient times, the uvula has been described as the source of diverse maladies, ranging from dehydration to viral and bacterial respiratory infections, as well as allergic reactions worsened by gastric refluxes. In Western Africa, traditional Africans Muslim – Hausa Fulani believe that once a child refuses mother's breast milk or presents with vomiting, diarrhea, anorexia, growth retardation or fever, he/she should undergo uvulectomy. This procedure, generally done by traditional barbers, is believed to facilitate swallowing, thus achieving normal growth and healthy living.³ In Nigeria, uvulectomy is commonly performed as a preventive measure against upper respiratory tract infection and chronic cough in neonates,⁴ in Morocco, as a mean to facilitate breast feeding and speech,⁵ and among Bedouins of south Sinai desert uvulectomy is considered as improving tolerance to thirst, and preventing upper respiratory tract diseases and pharyngeal obstruction.⁶

Uvular defects, as part of developmental abnormalities of the soft palate, may be associated with other facial midline defects, such as bifid uvula and clefts of the soft and hard palate. The presence of an elongated uvula (EU) is a quite rare malformation.

EU in childhood is usually asymptomatic, although it might cause obstructive apnea, especially in an infant with hypotonic oropharynx while lying supine, and may trigger gag reflex, regurgitation and reflex apnea.⁷ This abnormality, which could be also inherited, might account for such cases as the one presented here.

2. Case presentation

A five months old female, born after 25 weeks of gestation, was subsequently admitted to the neonatal intensive care unit for 20 weeks. Over the course of her hospitalization she suffered from necrotizing enterocolitis, complicated by perforation of the colon and obstructive jaundice, and retro-lental fibroplasia, both necessitating surgical interventions. The baby intermittently required assisted ventilation for short periods, and was treated with surfactant, steroids and diuretics. She had also developed septicemia on two occasions, and was treated with IV antibiotics.

Echocardiography performed during her hospitalization demonstrated a small cardiac ventricular-septal defect (VSD), which responded well to medicinal treatment. She was discharged home in good health, and a satisfactory growth and development, affected by episodes of cough. A week later, the child was found lying supine,

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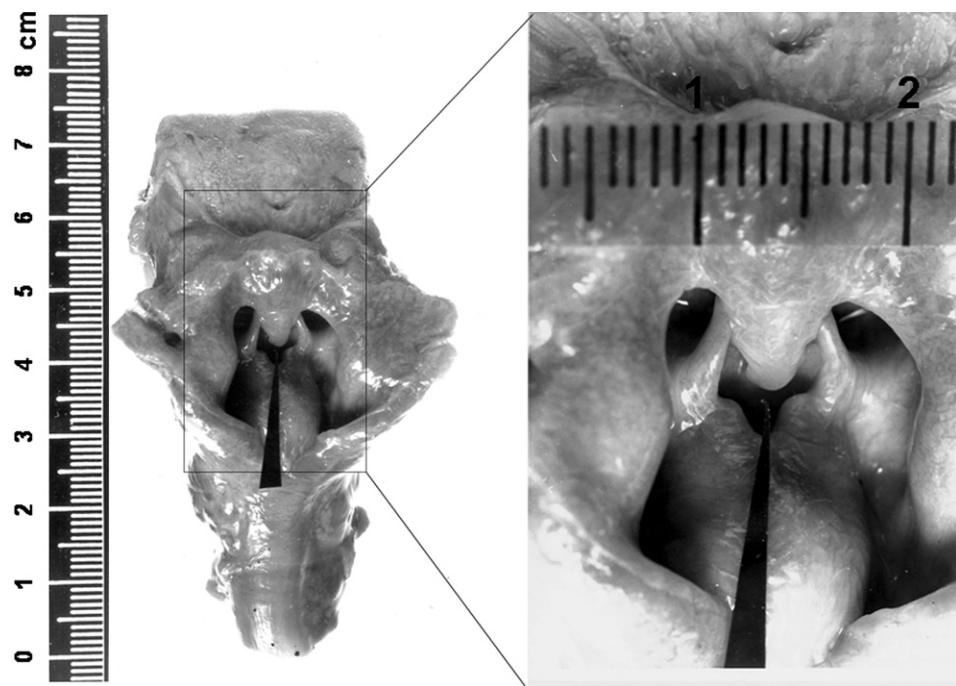


Fig. 1. Posterior view of the infant's pharynx at autopsy. Insert: an aberrantly EU (9 mm long) is impacted within the true vocal cords.

unconscious, in her crib. The body was delivered for examination at the National Center of Forensic Medicine under the provisional cause of death "sudden infant death syndrome" (SIDS).

At autopsy, the baby's developmental indices (weighted 2620 g, body length 49 cm) were those of a one-month-old female, with no external congenital anomalies. Yellowish mucus was present within the nostrils. Positive autopsy findings included a focal hemorrhage in the inferior lobe of the right lung, along with petechiae on the pericardium, thymus and visceral pleura. A small VSD of 0.5 cm and slight adhesions of the intraperitoneal organs were observed, probably as a consequence of repeated surgical interventions. An aberrantly long uvula, impacted within the true vocal cords, was found during the examination of the oral cavity (Fig. 1). The length of the uvula was 0.9 cm, its base 0.5 cm wide. There were no other gross pathological changes noticed.

The histological examination showed non-specific chronic inflammation of the uvula (Fig. 2), acute hepatic and splenic congestion, atelectatic foci in the lungs with local acute hemorrhages and without evidence of hyaline membrane disease (RDS). No evidence of intraperitoneal inflammation has been detected.

Bacteriology cultures from brain, lungs and heart tissues, blood and spinal fluid, and virology tests of blood, spinal fluid and nasal, oral and rectal swabs were all negative.

Due to the apparent lack of published data, and in order to assess the size of a normal baby's uvula, the uvula length of the seven consequent cases of infants, that were submitted to autopsy at our centre, were measured (Table 1). The average length of these babies' uvula amounted to 0.54 ± 0.05 cm.

3. Discussion

EU may protrude and come in contact with upper respiratory tract structures, such as the retropharynx, epiglottis and vocal cords. It can be the sole cause of chronic cough⁸ or contribute to its development, and should be suspected when no obvious reason for the cough is found on routine workup. Flexible bronchoscopy or fiberoptic examination are the preferred methods to evaluate the uvula, done at supine position.^{9,10}

A remarkably EU might cause more serious and life-threatening airway obstruction. Among other physical deformities, such as

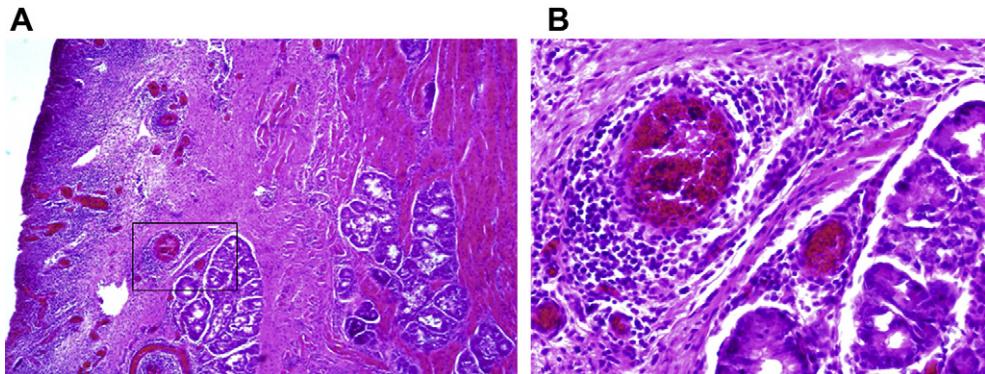


Fig. 2. Histology of the uvula. (A) General view of the EU. Note the presence of multiple salivary glands, as well as subepithelial inflammation (H&E, $\times 100$). (B) Higher magnification ($\times 400$) of the insert shown in (A). Non-specific perivascular chronic inflammatory infiltration is evident.

Table 1
Uvula length of 7 consecutive cases of infant death

Cause of death	Age (months)	Uvula length
Road traffic accident	3	0.6
Fall from height	2	0.53
Accidental hanging	4	0.46
SIDS	3	0.52
CO poisoning	6	0.61
Bilateral skull fracture	6	0.54
Fall from height	2	0.49
Average \pm S.D.	3.7 \pm 1.7	0.54 \pm 0.05

retrognathia and high hard palate, EU was prospectively found in as many as 15% of obstructive sleep apnea cases,¹¹ and may be a major contributor to its development. The presenting symptom might be an acute life-threatening event (ALTE), a well-known phenomenon concerning pediatricians, and the problem is sometimes solved only after surgical intervention.¹² The possibility that the uvula may cause apnea and SIDS was raised in the past,¹³ and a reported case of death attributed to this pathology exists.¹⁴

The above-described case was submitted for our investigation with the preliminary diagnosis of SIDS. Autopsy results suggested that the cause of death was asphyxia, ensued due to EU by either mechanical asphyxia due to obstruction of the glottis,¹⁵ or laryngospasm due to irritation of the vocal cords.¹⁶

It seems reasonable to assume that the longer than normal uvula descended between the vocal cords during sleep. At the end of expirium exhalation halts, and positive pressure necessary to dislodge the uvula from the glottis was absent. Choking may have wakened the baby abruptly, followed by a reflex attempt to inhale. The vocal cords, by action of the muscles, became rigid and tightened, trapping the large uvula (which by touching the larynx aggravated laryngospasm) thus causing further obstruction of the airflow. As the baby tried to inspire, the uvula was further sucked into the glottis, in a fatal vicious cycle. Considering that the glottis being the narrowest tract of the respiratory system, having it obstructed by the aberrantly large uvula, made the progression of asphyxia inevitable.

We propose that repeated intubations for assisted ventilation and general anesthesia during the infant's hospitalization must have rendered the uvula hypersensitive, and its aberrant size could be the result of reactive growth in response to recurrent irritation. Indeed, Haselby et al. showed that such mechanisms could be a reason for sudden stridor, airway obstruction and cyanosis.¹⁷ Laryngospasm in childhood could be associated with general anesthesia, gastroesophageal reflux disease, and foreign body aspiration.¹⁸ This condition is much more common among children than in adults, especially those younger than 12 months.¹⁹ Conclusively, trauma following upper respiratory tract instrumentation may cause EU.

It is also pertinent to note that the larynx of infants differs from that of adults mainly in the anatomic relation of neighboring structures. Neonates' epiglottis is located at the level of first cervical vertebra whereas the inferior border of the cricoid cartilage is located at the level of the fourth cervical vertebra. During growth of the pharynx, the epiglottis and cricoid cartilage descend to the levels of the 4th and 7th cervical vertebra, respectively. Therefore, infantile airway compromise can occur principally at rest, especially in supine position.

Although non-specific findings, such as petechiae of sclera, thymus or pericardium in the absence of other cause of death are consistent with SIDS which is still considered the major cause of death of babies younger than twelve months old, the presence of a malformation, such as EU, excludes SIDS.²⁰

The once obscure dilemma called SIDS, an entity in continuous debate,²¹ is not always the culprit of young babies' death, which can be simply the consequence of an anatomical mishap.

The presence of an unusually elongated uvula in an infant is suggested as the determining factor in this case for the fatal asphyxia. This rare case should be taken into consideration along with other congenital anomalies, such as bifid uvula and palatine clefts, in the differential diagnosis of a child presenting with episodes of cough or sleep apnea not otherwise explained. Post-mortem examination of infants suspected to succumb to SIDS or presenting findings compatible with asphyxia, such as petechiae of serosal surfaces of internal organs and foci of pulmonary atelectasis, should include morphometric evaluation of the respiratory tract.

Conflict of interest

None of the authors derives any support or other benefits from commercial sources reported on in the manuscript, no author has any other financial interest that could create a potential conflict of interest, or the appearance of a conflict of interest, with regard to this work.

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Ethical approval

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